Learning from errors

Recurrent attacks of headache and neck pain caused by congenital aplasia of the posterior arch of atlas in an adult

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Summary

A 47-year-old Chinese woman, with a history of recurrent attacks of vertigo and vomiting for the past 5 years, presented with intermittent radicular pain in the left upper limb for the past 2 years. She also reported recurrent attacks of severe headache and neck pain for more than 10 years. The pain might be aggravated by coughing or sneezing and relieved after sleeping in the decubitus position. The MRI depicted Chiari malformation. A multidetector CT scan and three-dimensional CT reconstruction revealed partial aplasia of the left posterior arch of atlas of a small gap. The patient underwent plastic surgeries in Beijing. The disappearance of the recurrent pain syndrome was confirmed by follow-up after surgery.

BACKGROUND

Chiari malformation and basilar invagination belong to the anomalies of the craniovertebral junction, which are commonly considered to be childhood or adolescent syndromes.¹ Paediatricians and orthopaedics are familiar with these disorders. In the present case, we describe an adult patient with acquired Chiari malformation and basilar invagination caused by congenital aplasia of the posterior arch of atlas, who suffered from recurrent attacks of headache and neck pain for more than 10 years, and had been misdiagnosed for several years.

It has been reported that congenital aplasia of the posterior arch of atlas may increase the risk of atlantoaxial subluxation in about 26% of children aged 2–3 years.² The stability of atlantoaxis in adolescence may be due to the connective tissue bridging the small bony defect, which was found in autopsies or operations.³ Due to the progress of the atlantoaxial instability, our patient developed paroxysmal obvious symptoms as a result of compression of the brain stem, cerebellum and the roots of cervical nerve.

This case sheds a new light on the anomalies of the craniovertebral junction like basilar invagination as a possible cause of headache and neck pain in adult patients. It is important, especially for radiologists and physicians, to keep in mind the possibility of basilar invagination and the related deformities.

CASE PRESENTATION

A 47-year-old Chinese woman, with a history of recurrent attacks of vertigo and vomiting for the past 5 years, presented with intermittent radicular pain in the left upper limb for the past 2 years. She also reported recurrent attacks of severe headache and neck pain for more than 10 years. The pain might be aggravated by coughing or sneezing and relieved after sleeping in decubitus position. The patient had presented to local hospitals for episodes of headache

and sudden vertigo many times. No abnormal signs were documented in the previous medical records. A brain MRI scan was suggested (figure 1A). The report showed ‘no obvious brain abnormalities’. Considering the diagnosis of ‘tension-type headache’ and ‘Meniere’s syndrome’, the patient was treated conservatively with oral analgesia and betahistine but similar symptoms still occurred from time to time.

Neurological examination was normal and the result from the examination of cranial nerves was completely normal. No motor deficit or sensory loss was revealed and reflexes were brisk bilaterally. The Hoffmann, Romberg, Babinski and Lhermitte signs were negative.

INVESTIGATIONS

The patient had a MRI of her cervical spine after complaints of radicular pain in the left arm. The MRI (figure 1B) depicted Chiari malformation (downward hernia of the cerebellar tonsils and the medial portions of the inferior cerebellar lobes). The brain stem seemed to be compressed by the odontoid process. A multidetector CT scan and three-dimensional CT reconstruction revealed partial aplasia of the left posterior arch of atlas (figure 1C–I) and underlying basilar invagination (figure 1J–L). It is worth noting that the previous brain MRI data review also found the Chiari malformation (figure 1A).

DIFFERENTIAL DIAGNOSIS

The terms ‘Chiari malformation’ and ‘basilar invagination’ are interrelated but different. The former refers to hind-brain herniation and the latter is defined as a radiographical finding of anomalies in the craniocervical junction rather than a clinical diagnosis itself. Both Chiari malformation and basilar invagination may be congenital or acquired. Our patient was a case of acquired Chiari malformation and basilar invagination caused by congenital aplasia of the posterior arch of atlas.

Figure 1 Mid-sagittal T2-weighted MR image (A, B). Multidetector CT scan (C–F). Note the incomplete left posterior atlas arch in D and F. The right posterior arch of atlas is intact in C and E. Three-dimensional CT reconstruction (G–I). Note the partial aplasia of the left posterior atlas arch in G and I. CT reconstructed images (J–L).

TREATMENT

The patient underwent a number of operations in Beijing, including posterior cranial fossa decompression, cisterna magna enlargement neoplasty, posterior atlantoaxial lateral mass screw-plate fixation and fusion.

OUTCOME AND FOLLOW-UP

The disappearance of the recurrent pain syndrome was confirmed by follow-ups after surgery.

DISCUSSION

Chiari malformations were first described by pathologist Hans Van Chiari in 1891 and classified as types I–IV (table 1).⁴ Our patient had a type I Chiari malformation (CIM). The CIM consists of caudal displacement of the cerebellar tonsils at least 3 mm into the upper cervical spinal canal.⁵ Patients with the CIM may present with a variety of symptoms and signs ranging from a slight headache to severe myelopathy and brainstem compromise. The most

Figure 2 Classification of posterior arch defects of the atlas. Type A: failure of posterior midline fusion with a small gap remaining. Type B: unilateral clefts. Defects may range from a small gap to a complete absence of the hemi-arch. Type C: bilateral defects with preservation of the most dorsal part of the arch. Type D: complete absence of the posterior arch with a persistent isolated tubercle. Type E: complete absence of the entire posterior arch, including the posterior tubercle. Over 97% of defects are type A. The incidence of the general population harbouring types B–E was estimated 0.69%.³

Table 1 Types of Chiari malformations

Chiari type	Anatomical description
Type I	Herniation of the cerebellar tonsils and sometimes part of the medulla below the foramen magnum often associated with a 'too small posterior fossa'
Type II	Herniation of the cerebellar vermis, brain stem and fourth ventricle below the foramen magnum. Typically associated with myelomeningocele and hydrocephalus. Type II Chiari malformations are sometimes referred to as the Arnold-Chiari malformation
Type III	Herniation of the entire posterior fossa contents and brain stem into a cervical encephalocele
Type IV	Cerebellar hypoplasia

Cited from Speer *et al.*⁴

common symptom is pain, usually occipital and upper cervical in location, and often induced or exacerbated by Valsalva manoeuvres such as laughing, sneezing and coughing, which may provoke dynamic cord compression. Our patient had similar attacks of headache and neck pain for more than 10 years.

Basilar invagination refers to a developmental anomaly in which the vertebral column is situated in an abnormally high position due to decreased height of the skull base. The odontoid process becomes higher than the normal level into the foramen magnum. The diameter of foramen

magnum shortens and the volume of posterior fossa becomes smaller, following with the hindbrain herniation and the compression of medulla oblongata, cerebellum and cervical nerve roots. Basilar invagination may be caused by congenital or acquired anomalies of occiput, atlas and axis, as well as acquired skull-base softening. The tip of the odontoid process projects more than 5 mm above Chamberlain's palato-occipital line.⁶ Ventral spinal cord compression occurs with Wackenheim's clivus-canal angle less than 150°. The atlanto-occipital joint axis angle is more obtuse than the average range (124–127°).⁶ The tip of the odontoid process was 4.7 mm above Chamberlain's line in our patient (figure 1J). Nevertheless, both of the angles reached the standards: the clivus-canal angle was 126.86° (figure 1K) and the atlanto-occipital joint axis angle was 146.62° in this case (figure 1L).

The clinical presentation of basilar invagination is diverse too. A constellation of symptoms and signs that reflect dysfunction of the brain stem, cerebellum, cervical spinal cord, upper cervical nerve roots, lower cranial nerves and the vascular supply point to the craniocervical border.⁶ The main symptoms in this case were recurrent attacks of headache, vertigo and cervical radicular pain.

Total or partial aplasia of the posterior arch of atlas is relatively uncommon. Geipel^{7–10} found it in 4% of 1613

autopsies. The posterior arch of the atlas begins its ossification at the seventh week of intrauterine life.¹¹ This ossification process starts from two centres in the lateral masses and proceeds to the midline.¹² At birth, the posterior arches are nearly fused except for several millimeters of cartilage, while the complete ossification is expected to occur between 3 and 5 years of age. Congenital aplasia of the posterior arch of atlas is thought to be due to a failure of the normal posterior arch chondrification process. Currarino *et al*¹³ developed classification types (A–E) for congenital aplasia of the posterior arch of atlas (figure 2), 97% of which were type A. The incidence of the general population harbouring types B–E was estimated 0.69%. Our patient had a type B anomaly, which was a small gap.

This case illustrates the lack of recognition of Chiari malformation and basilar invagination in adult patients. It is important, especially for radiologists and physicians, to keep in mind the possibility of basilar invagination and the related deformities.

Learning points

- ▶ This case illustrates the lack of recognition of Chiari malformation and basilar invagination in adult patients.
- ▶ It is important, especially for radiologists and physicians, to keep in mind the possibility of basilar invagination and the related deformities.
- ▶ The stability of atlantoaxis in adolescence may be due to the connective tissue bridging the small bony defect, which was found in autopsies and operations.

Competing interests None.

Patient consent Obtained.

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